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Laryngomalacia

Introduction:

The term Malakia in Greek ¹ means softening. The term indicates softening of larynx.

This is a disease of infants and children. This disease is characterised by the presence of stridor (inspiratory) which is caused due to excessive redundancy of supraglottic tissues ² which gets sucked into the glottis due to the negative pressure caused during inspiration. The term laryngomalacia was first coined by Chevalier Jackson in 1942 ³. The stridor in these patients are inspiratory in nature. Laryngomalacia is the leading cause of stridor in infants accounting for approximately 70% ⁴.

Age of onset:

This condition gets manifested within first two weeks after birth and usually resolves when the child reaches the age of 2.

Pathophysiology ⁵:

1. Excessive redundancy of supraglottic soft tissues
2. Immaturity of neuromuscular system
3. Immaturity of cartilages of larynx
4. GERD
5. Submucosal gland hyperplasia

Possible mechanisms causing stridor in these patients include:

1. Indrawing of cuneiform cartilages on inspiration
2. Omega shaped epiglottis of infants curls upon itself
3. The arytenoids collapse inwards
4. The epiglottis gets displaced against the posterior pharyngeal wall
5. Short aryepiglottic folds
6. Overly acute angle of epiglottis at the laryngeal inlet

Clinical features:

1. These patients have stridor which is inspiratory in nature is present immediately after birth.
2. Stridor is intermittent, position and becomes worse on activity
3. Stridor is usually preceded by an episode of upper respiratory tract infection
4. These children may have sternal recession which may lead to pectus excavatum and cor pulmonale
5. These children may have feeding difficulties
6. GERD may also be associated

Types of laryngomalacia:

Type I: Inward collapse of aryepiglottic folds

Type II: Long tubular epiglottis infolding on itself (Omega shaped)

Type III: Anterior and medial collapse of corniculate and cuneiform cartilages

Diagnosis:

Type IV: Posterior displacement of epiglottis against posterior pharyngeal wall or inferior collapse towards vocal folds

Type V: Short aryepiglottic folds



Type I laryngomalacia



Type II laryngomalacia



Type III laryngomalacia



Type IV laryngomalacia



Type V laryngomalacia

Is usually made by performing direct examination of larynx under anesthesia. It is not possible to perform indirect laryngoscopy using a laryngeal mirror in these infants. Flexible nasopharyngoscope is used to visualize larynx in these infants. This is usually performed under general anesthesia. These infants classically show inward collapse of supraglottic structures the moment suction tip is placed at the level of vocal cords. This classic sign is known as "Narcy sign". This sign is pathognomonic of laryngomalacia.

During laryngoscopy three anatomical anomalies existing either separately or in combination can be identified. They include:

1. Flaccid epiglottis
2. Poorly supported arytenoids
3. Short aryepiglottic folds

It has been suggested that laryngeal examination be performed in an awake infant in order to study the mobility of laryngeal structures in real time. This procedure has its own pitfalls which include:

1. Vocal cord abduction as a result of crying may mask the presence of laryngomalacia
2. Splinting effect of laryngoscopy blade at the level of vallecula may prevent effectively the collapse of supraglottic structures.

Management:

Majority of infants with laryngomalacia recover spontaneously as their cartilages and neuromuscular system matures. They recover completely by the time they reach the age of 2. Only supportive measures need to be administered in order to alleviate the agony of the suffering infant. As soon as the acute crises is tided over they undergo spontaneous recovery.

Surgery:

Is indicated only in patients with severe disease. Indications for surgery include:

1. Dyspnoea / severe on exertion
2. Feeding difficulties
3. Failure to thrive
4. Sleep apnoea
5. Obstructive hypoventilation
6. Cyanosis
7. Bradycardia
8. Uncontrollable reflux

Prior to 1980 tracheostomy was performed in order to alleviate symptoms. Currently supraglottoplasty is the treatment of choice. The major advantage of this procedure is that it addresses the area of obstruction directly. It is advisable to perform unilateral supraglottoplasty initially.

Types of supraglottoplasty:

1. Microlaryngeal supraglottoplasty
2. Laser assisted supraglottoplasty
3. Microdebrider assisted supraglottoplasty

Direct laryngoscopy / bronchoscopy should be performed prior to surgical management because of other associated synchronous airway lesions. While performing direct laryngoscopy the following factors should be noted:

1. Air way oedema
2. Erythema
3. Anatomical structure of larynx
4. Laryngeal movement
5. Rotation of epiglottis posteriorly and arytenoids anteriorly
6. Areas of inward collapse of supraglottis is identified
7. Static end points of inspiration and expiration

History of supraglottoplasty:

In 1922 Iglaue first described endoscopic removal of supraglottic tissue using a nasal snare.

In 1984 Dr Lane described removal of corniculate cartilage along with redundant interarytenoid mucosa.

In 1985 Dr Seid described the use of carbondioxide laser in the management of laryngomalacia.

Surgical procedure:

Topical xylocaine 4% without adrenaline is sprayed over the laryngeal mucosa in order to anesthetize the area. This procedure reduces adduction reflex of the vocal folds during direct laryngeal examination. Based on the areas of anatomic narrowing a definitive surgical modality is planned.

The tip of the suspension laryngoscope used for surgery should be placed in the vallecula or against the laryngeal surface of epiglottis. The redundant mucosa over the aryepiglottic folds can be excised by using microlaryngeal instruments. If necessary the whole of aryepiglottic fold on one side can be sectioned. It is better to perform this surgery on one side, and if the effects are not satisfactory the aryepiglottic fold on the opposite side should be attended to at a different sitting.

Powered instruments like microdebrider can be used to perform supraglottoplasty. After creating a raw area then a debrider blade is used to excise the redundant mucosa. Laser can also be used to perform supraglottoplasty.

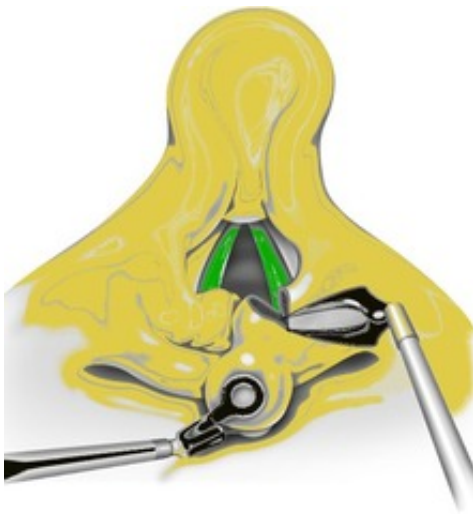


Image showing supraglottoplasty procedure

Complications:

1. Wound sepsis
2. Granulation tissue formation
3. Supraglottic stenosis (difficult to manage)

Aryepiglottoplasty:

This procedure was first reported in 1984. This procedure involves resection of lateral portion of epiglottis, corniculate cartilages and the tips of arytenoids.

Aryepiglottic fold division:

In this procedure aryepiglottic fold is divided. This prevents infolding of the aryepiglottic folds during inspiration.

Microdebrider assisted supraglottoplasty:

This method was popularized by Zalzal in 2005. He used a microdebrider to perform supraglottoplasty.

1. Aryepiglottic folds are resected using a microdebrider - Anteriorly up to the epiglottis and posteriorly up to the arytenoid cartilages
2. Redundant supra arytenoid mucosa is removed using a debrider

Conclusion:

Majority of laryngomalacia patients improve with conservative management and reassurance. Majority of these children outgrow their problem. Rarely surgical intervention were resorted to. In all these patients coexisting lesions like GERD should be ruled out.

Video clipping demonstrating videolaryngoscopy being performed on a baby with laryngomalacia

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